British Heart Journal, 1978, 40, 931-933

Systemic sarcoidosis with refractory ventricular tachycardia and heart failure

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SUMMARY A patient who presented with recurrent arrhythmias, including refractory ventricular tachycardia, and heart failure, was later found to have sarcoidosis with pulmonary and lymph node involvement.

Myocardial sarcoidosis was once believed to be quite rare and usually diagnosed only at necropsy but is now more often diagnosed and treated in life. In a small number of patients dramatic remission of the most severe manifestations of sarcoid heart disease has followed the institution of steroid therapy. In the case reported here, there was clinical, radiographic, and histological evidence of systemic sarcoidosis; though histological confirmation of involvement of the heart by sarcoidosis is lacking, the clinical manifestations, the serious and widespread rhythm disturbances, the similarity of this case to some other reported cases (Duvernoy and Garcia, 1971), and the response to steroid therapy are strong evidence in favour of myocardial involvement by the granulomatous process.

Case report

On 29 May 1975, a 36-year-old white man presented with a history of two episodes of dyspnoea, palpitation, and anterior chest pain, each lasting 10 minutes and occurring 48 hours before admission. One year before, acute iritis had been diagnosed. Six months before admission, he developed anorexia, fatigue, and polyarthralgia, together with a cough productive of white phlegm. He had no other symptoms and there was nothing of note in the family, social, or dietary history.

He was a well-developed man, with no fever, rash, nodules, lymphadenopathy, icterus, clubbing, or cyanosis. Blood pressure was 110/70 mmHg. The pulse was 80/minute and regular. The jugular veins were distended to the angle of the mandible at 45°. The apical impulse was displaced to the 6th left intercostal space midway between the midclavicular and the anterior axillary line, and there was a slight left parasternal heave. The first sound was normal, the second sound was normally split with accentuated pulmonary component, and a fourth heart sound was heard. The remainder of the physical examination was normal.

The electrocardiogram showed frequent multifocal ventricular premature beats and occasional atrial ectopic beats; there were widespread nonspecific ST segment and T wave changes. Chest *x*-ray film showed a cardiothoracic ratio of 18/34, the proximal pulmonary vessels were prominent and there were bilateral basal interlobular septal lines. CK and AST levels were normal. Haemoglobin, red cell indices, white cell count and differential, and platelets were all normal. The following additional investigations gave negative or normal results: serum bilirubin, alkaline phosphatase, calcium and inorganic phosphorus, antistreptolysin titre, antinuclear factor, LE cells, rickettsial and viral serology, toxoplasma dye test, serum iron and iron-binding capacity, and blood cultures.

Initially, the patient was treated with digoxin and quinidine but the atrial and ventricular ectopic beats persisted. Some weeks after admission, tightness in the chest and palpitation recurred and the electrocardiogram showed a supraventricular tachycardia with frequent multifocal ventricular ectopic beats still present. After intravenous practolol the blood pressure fell, but with infusions of lignocaine, phenytoin, and hydrocortisone, sinus rhythm returned and the blood pressure rose to normal. Twenty-four hours later he developed signs of acute heart failure, with pulmonary oedema on the chest *x*-ray film, and was treated with frusemide. Ventricular tachycardia then developed

despite continued quinidine treatment, but was successfully treated by lignocaine infusion and procainamide. During the subsequent days, ventricular tachycardia recurred frequently and required electrical countershock on 30 or more occasions together with lignocaine, quinidine, procainamide, and phenytoin. During an episode of cardiorespiratory arrest he was intubated and given intermittent positive pressure ventilation, but remained unconscious for 5 hours. Ten weeks after admission the patient was symptomatically well despite persistence of atrial and ventricular ectopic beats. He was discharged home on treatment with frusemide and procainamide without an aetiological diagnosis.

A follow-up chest *x*-ray film was taken on the 9 September 1975; the heart size was now normal, with a cardiothoracic ratio of 15/34, and the pulmonary oedema present earlier had completely cleared; there was however bilateral hilar node enlargement with fine nodulation and reticulation, particularly at the right base and in both midzones. Hilar node enlargement was confirmed by tomography. It is possible that these *x*-ray changes suggesting sarcoidosis were present on previous films but were obscured by the signs of pulmonary oedema. The patient was readmitted for liver biopsy and mediastinoscopy. On this occasion, enlarged lymph nodes were noted in the left axilla and left supraclavicular region; biopsy showed near total nodal replacement by multiple noncaseating granulomas consistent with sarcoidosis. Pulmonary function testing showed normal lung volumes and flow rates and the transfer factor was normal.

When the diagnosis of sarcoidosis was confirmed, the patient was started on treatment with prednisone 60 mg daily. Isoniazid was also given because of a positive Mantoux reaction. Frusemide and procainamide were continued. He has now remained in good health for the 12 months since starting treatment with corticosteroids; ventricular tachycardia and pulmonary oedema have not recurred. The most recent electrocardiograms show an impressive reduction in ventricular ectopic activity with long periods of uninterrupted sinus rhythm. In addition, the chest *x*-ray appearances are now normal.

Discussion

Cardiac involvement in a patient with sarcoidosis was first observed by Bernstein *et al.* in 1929 and subsequently by Nickerson in 1937. In 1960, Porter reviewed earlier reports and found 33 documented cases of sarcoid heart disease with necropsy confirmation; he reviewed an additional 20 cases with cardiac involvement where death may have been the result of other causes. In 1971, Gozo *et al.* reported two new cases and collected from the entire world literature a total of 70 fatal cases. Reports of this disease in Britain have been rare, but in 1974 Fleming collected and reported 50 new cases, 20 of which were fatal and had been confirmed at necropsy.

Granulomatous infiltration of the heart in sarcoidosis can affect the heart in a number of ways. The most frequent manifestations include conduction disturbances, heart failure, and sudden death. Infrequent manifestations include mitral regurgitation from fibrous replacement of papillary muscles, left ventricular aneurysm, pericardial effusion, mural thrombi, haemorrhagic pericarditis, and electrocardiographic patterns of transmural infarction. Sarcoid granulomas have been described in the aorta, the superior vena cava, and the pulmonary veins. The typical presentation, however, is with conduction system abnormalities, most often atrioventricular or intraventricular conduction defects, or disorders of impulse formation, including atrial tachycardia, ventricular ectopic beats, and ventricular tachycardia. In addition, non-specific changes on electrocardiogram have been found in approximately 40 per cent of patients. Adams-Stokes syndrome and sudden death are the most serious consequences of these rhythm disturbances.

Heart failure is less common and may be a manifestation of a rhythm disturbance, widespread fibrosis resulting from granulomatous infiltration of the myocardium, left ventricular aneurysm, or rarely cor pulmonale secondary to pulmonary fibrosis and pulmonary hypertension, or a combination of any of these processes. Our patient developed many of the reported arrhythmias and during his time in hospital was seen to have supraventricular tachycardia, atrial ectopic beats, multifocal premature ventricular beats, atrial fibrillation and flutter, junctional tachycardia, and recurrent ventricular tachycardia. The refractoriness of the ventricular arrhythmias noted in our patient appears to be characteristic of this disorder (Stein *et al.*, 1962; Bashour *et al.*, 1968).

Various authors have reported benefits from the use of corticosteroid therapy for patients with electrocardiographic or clinical evidence of involvement of the heart by sarcoidosis. The relative rarity of serious cardiac dysfunction in patients with systemic sarcoidosis and the obvious reluctance to withhold treatment which might be beneficial or even lifesaving has made controlled trials impossible and there are no reports on the use of other drugs effective in sarcoidosis being used either as supplements to corticosteroids or as alternatives to

steroids in this condition. In our patient it is hoped at least that steroids will halt the progression of the disease and that the various rhythm disturbances will be controlled by continuing conventional antiarrhythmic drug treatment.

I am indebted to Professor C. F. McCarthy, University College and Regional Hospital, for permission to report on this patient under his care, and to him and Dr P. Finnegan, Regional Hospital, Galway, for advice and encouragement in the preparation of this paper.

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